CASE REPORT

A rare presentation of paracentral acute middle maculopathy during early gestation

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SUMMARY

Paracentral acute middle maculopathy (PAMM) is a rare ischemic pathology affecting the macula, specifically the middle retinal layers. Patients often present with an acute onset of paracentral scotoma, but some may be asymptomatic. It is most often seen in patients with vascular risk factors or retinal vascular occlusive disease. PAMM is exceedingly rare in pregnancy and is commonly overlooked during clinical examination. Diagnosis is usually made with the support of Optical Coherence Tomography (OCT) or Optical Coherence Tomography Angiography (OCTA) imaging. We report a case of PAMM in a young, healthy pregnant woman who complained of sudden onset of unilateral paracentral scotoma and exhibited imaging features of PAMM. Her systemic workup was unremarkable, and she was managed conservatively with close monitoring. She achieved complete resolution by seven weeks postpartum. This case emphasizes the clinical relevance of considering PAMM in the causes for pregnant patients presenting with acute visual symptoms. The combined use of OCT and OCTA is key in identifying PAMM, especially when funduscopic findings are subtle and other vascular retinopathies are not evident.

INTRODUCTION

Paracentral acute middle maculopathy (PAMM) was first reported in 2013 by Sarraf et al, identifying it as a subdivision of acute macular neuroretinopathy (AMN).[1] Advances in spectral-domain Optical Coherence Tomography (SD-OCT) have greatly improved the ability to localize lesions in AMN. PAMM is considered an uncommon retinal vascular condition, typically identified on SD-OCT as a hyperreflective band at the level of the inner nuclear layer (INL).[2] The typical clinical presentation of PAMM is an acute paracentral scotoma with preserved central acuity. Since fundus examination usually reveals only subtle or non-specific changes, Optical Coherence Tomography (OCT) and Optical Coherence Tomography Angiography (OCTA) are often required to establish the diagnosis.[3] However, access to these imaging is often limited, thus leading to the condition being underdiagnosed in routine practice.

Although the pathogenesis of PAMM remains incompletely understood, several factors have been identified, which include systemic or pharmacologic vasoconstrictors, such as adrenaline and caffeine, as well as hormonal influences like oral contraceptive use. Moreover, retinal vascular causes include diabetic retinopathy, central retinal vein occlusion, branch retinal artery occlusion and sickle cell retinopathy.[2] Reports of PAMM during pregnancy are uncommon, with proposed mechanisms including a hypercoagulable state of pregnancy, maternal anaemia, and systemic hypotension, all of which may contribute to retinal ischaemia. Given its non-specific presentation and rarity in pregnancy, we report a case of PAMM occurring in early gestation in a healthy young woman without vascular risk factors, emphasizing the clinical and imaging features that facilitated the diagnosis.

CASE PRESENTATION

A healthy 33-year-old primigravida, at 10 weeks of gestation, with no prior medical illness, complained of right temporal paracentral scotoma for one week. The symptom was described as a persistent "glitch of light" obscuring her visual field. She otherwise had no other ocular or neurological symptoms. She was normotensive throughout pregnancy. On examination, visual acuity was bilaterally 6/6, with no relative afferent pupil defect. The anterior segment examination revealed normal findings with intraocular pressure of 14mmHg bilaterally. Funduscopy examination of the right eye showed a slight elevation nasal to the fovea. The left fundus appeared normal. There was an enlarged blind spot in the right Humphrey visual field (HVF) test (Figure 1).

Meanwhile, the right OCT of the macula showed a hyperreflective band nasal to the fovea, confined to the INL, findings characteristic of PAMM (Figure 2a). OCT angiography (OCTA) detected a diminished flow at the site of the lesion (Figure 2b). Systemic investigations, including blood pressure monitoring, fasting blood sugar, lipid profile, echocardiography, and carotid ultrasound Doppler, showed no abnormalities. Thus, a diagnosis of PAMM was derived, and she was followed up closely. During follow-up, her scotoma gradually improved, accompanied by a progressive reduction in the lesion on OCT. At seven weeks postpartum, the patient experienced minimal residual scotoma, with complete resolution of the hyperreflective band and normal OCTA (Figure 3).

DISCUSSION

Acute onset of visual disturbance during pregnancy is concerning as it may reflect a potentially serious underlying

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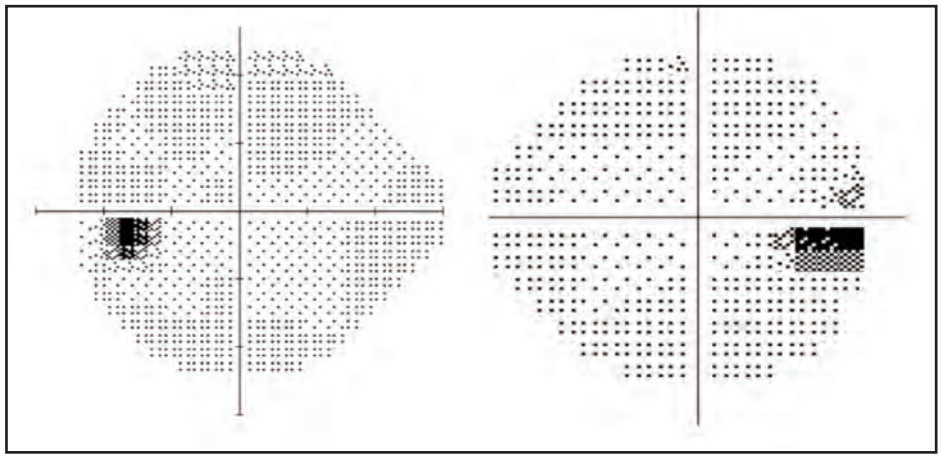

Fig. 1: HVF* of the right eye shows an enlarged blind spot on presentation, whereas the left eye Humphrey Visual Field appears normal.

*HVF, Humphrey Visual Field.

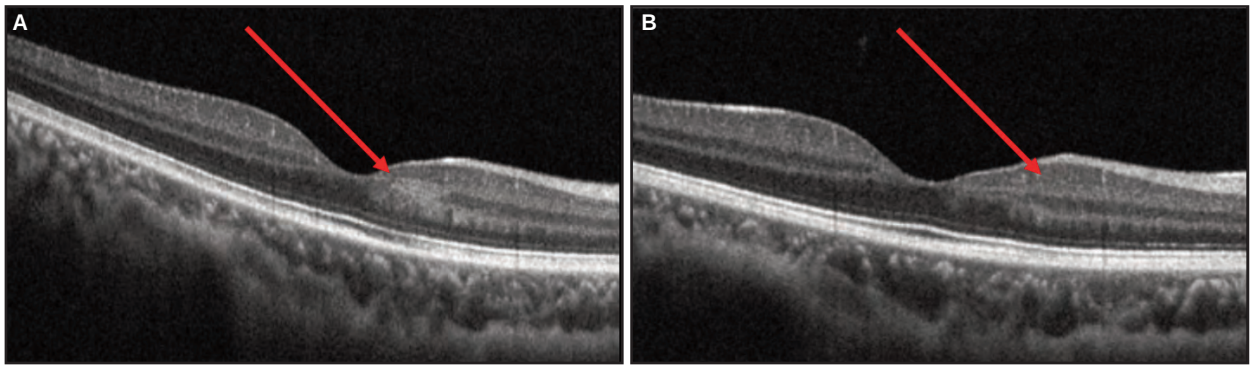

Fig. 2: A) OCT† of the right macula revealed a hyperreflective band nasal to the fovea, which involved INL‡ on presentation. B) At seven weeks postpartum, OCT† of the macula demonstrated the disappearance of the previously noted nasal parafoveal hyperreflective band, without evidence of retinal layer thinning.

†OCT, Optical Coherence Tomography; ‡INL, inner nuclear layer.

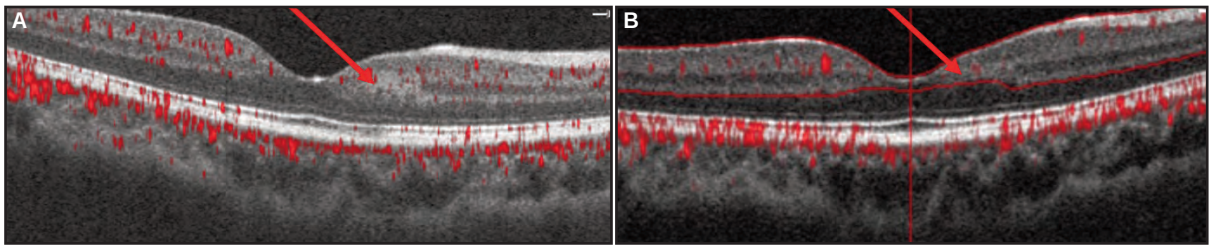

Fig. 3: A) OCTA§ of macula revealed flow attenuation that matched the location of the lesion at initial presentation. B) No more abnormality seen on OCTA§ of the macula.

§OCTA, Optical Coherence Tomography Angiography.

pathology. Such scenarios frequently impose a considerable dilemma for clinicians in both diagnosis and management. Visual impairment in pregnancy often happens, resulting from severe hypertensive complications or the physiologic hypercoagulable state, both of which lead to a significant risk to ocular wellbeing.[4] PAMM occurring during pregnancy is highly unusual. Typical cases of PAMM are usually associated with older age, underlying vascular risk factors, systemic diseases, or retinal arterial occlusive diseases. In contrast, our patient was young and otherwise healthy, with pregnancy identified as the only plausible contributing factor.[2]

To date, there have only been two reported cases of PAMM occurring during pregnancy. It was first documented by Pencen et al., who involved a 29-year-old pregnant woman with no known medical illness, who developed a paracentral scotoma during a presyncope hypotensive episode. OCTA showed evidence of retinal infarction, revealing a persistent reduction in deep retinal capillary perfusion with subsequent atrophy of the INL. It was postulated that a combination of hypotension, anaemia, and the hypercoagulability that happens during pregnancy may have contributed to the development of PAMM in this patient.[5]

Another author proposed that the mechanism behind PAMM involves an imbalance in retinal oxygen dynamics. The reduced blood flow velocity and lower oxygen saturation at the venular pole lead to increased oxygen extraction from the retinal arterial circulation, resulting in ischemic damage to the middle retinal layers. This is evidenced by findings consistent with OCTA that showed disruption of the deep capillary plexus and decreased vascular density in the superficial capillary plexus, indicating a more severe degree of microvascular compromise in PAMM.[6]

Nevertheless, PAMM does not directly lead to these changes, as the superficial plexus operates independently from the deep retinal circulation. Instead, this pattern likely results from a shared systemic cause, such as hypertension, as simultaneous microvascular alterations occur throughout the retina.

PAMM typically presents with a sudden paracentral scotoma, which may be accompanied by blurred vision. PAMM lesions are situated in the deeper parafoveal retina and appear grayish white, which is distinguished from cotton wool spots. However, due to their minimal visibility, these lesions are often missed during routine fundus examinations and are more reliably visualized with SD-OCT using near-infrared reluctance imaging.[3]

Type 1 PAMM is marked by the involvement of retinal structures above the outer plexiform layer (OPL), which is likely corresponding to vascular compromise in the superficial or intermediate capillary plexus.

The characteristic feature of PAMM is the presence of a hyperreflective band extending across the middle retinal layers. PAMM can be divided into two patterns based on the location of the lesion. Type 1 involves the superficial or intermediate capillary plexus, affecting retinal layers above the OPL. In contrast, Type 2 affects the layers beneath OPL, consistent with ischaemia originating from the deep capillary plexus.[1]

With the emergence of OCTA, the underlying pathophysiology of PAMM is now better understood. Recent studies suggest that hypoperfusion of the deep capillary plexus is a key feature during the acute stage of the disease. This helps explain the pattern of selective INL injury seen in acute PAMM and the ongoing thinning of this layer in the chronic stage.[7] OCTA has become essential for differentiating Type 1 and Type 2 PAMM, as its imaging findings reveal ischemic damage at the intermediate and deep capillary plexus levels, accounting for the characteristic lesion patterns observed.[3]

Clinical reports have linked symptomatic PAMM to a variety of pharmacological agents, as well as numerous ocular and systemic disorders. Recent findings indicate that small lesions, although sharing the typical morphology of PAMM, often occur without symptoms and are typically diagnosed retrospectively, after the lesion has resolved.[6] These small PAMM lesions frequently appear in patients with retinal vein occlusions, hypertension, diabetes (with or without retinopathy), underlying cardiovascular disease, or elevated vascular risk.[8] Various ocular and systemic conditions have been linked to PAMM through their effects on the retinal circulation. Examples include compressive ocular ischemia, hematological disorders such as sickle cell disease, trauma related entities such as Purtscher's retinopathy, inflammatory vasculitis, and systemic conditions including uncontrolled hypertension, migraine, and even viral upper respiratory infections. The diversity of underlying causes supports the idea that PAMM should be reviewed not as a distinct disease, but rather as a manifestation of retinal vascular compromise reflecting broader systemic or ocular circulatory dysfunction. In this regard, PAMM could serve as a clinical marker of vascular morbidity or as an indicator within the spectrum of retinal vascular disorders.[9]

In distinguishing PAMM from optic neuritis, standard clinical tests such as visual acuity, color vision, RAPD, and visual field assessment are useful. However, if results are inconclusive, clinicians should consider a retinal cause as a potential explanation. In practice, OCT and OCTA are highly valuable for confirming whether the optic nerve or retina is involved, helping to prevent unnecessary investigations- a particularly important factor during early pregnancy. Moreover, OCTA provides insights into the vascular pathophysiology of PAMM.

Although PAMM lesions typically resolve over time, most patients continue to experience scotomas. As the condition evolves, OCT imaging reveals that the hyperreflective band usually disappears within a few months, often followed by thinning and distortion of the INL and OPL of the middle retina.[1] Additional changes may include excavation of the inner retinal surface and thickening of the ONL.[10] After the lesion resolves, OCTA often shows areas of capillary dropout and structural disruption of the deep capillary plexus,

highlighting the chronic vascular consequences of PAMM.[10] Fundus fluorescein angiography generally does not provide further detail about PAMM lesions but remains helpful in excluding ischemic conditions related to other disorders.

PAMM may be the only ocular indicator of an underlying systemic microvascular disorder, even in patients with no obvious comorbidities. As a result, a thorough systemic evaluation is essential to rule out possible cardiovascular risk factors. Early identification and prompt treatment can help reduce mortality. However, when assessing young individuals with sudden unilateral visual impairment and normal fundus, clinicians must also consider the possibility of an underlying retinal disorder.

CONCLUSION

Although uncommon, PAMM should be considered as an important cause of paracentral scotoma even in healthy pregnant women without known vascular risk factors. OCT and OCTA are critical for early detection, monitoring disease progression, and excluding other possible causes. Although no specific treatment is available, prompt recognition of PAMM and evaluation of systemic risk factors, particularly cardiovascular and hematological abnormalities, are essential for optimal patient management.

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